Neuromyelitis Optica as a Possible Cause of Severe Retrobulbar Optic Neuritis
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Neuromyelitis optica (NMO) is a rare demyelinating disease presenting with optic neuritis and spinal cord involvement, with less brain involvement than in multiple sclerosis (MS). Visual prognosis and long-term treatment also differ; therefore accurate diagnosis is essential.

1. Case History

A 57 year old African-American female presented with a chief complaint of sudden vision loss OD which began two days prior. Upon questioning she admitted to mild eye pain with movement OD and intermittent tingling of her left hand over the past year. The patient's medical history included hypertension for 25 years, borderline diabetes, hepatitis C, and a cancerous kidney cyst which was removed in 2007. Her medications included Nifedical, atenolol, furosemide, methimazole, lisinopril, and penicillin for a possible tooth infection. Her social included a 20 year history of smoking one pack per day.

2. Pertinent Findings

At initial presentation, best-corrected vision measured 20/100 through eccentric viewing OD and 20/20 OS. Pupils were equal and round with a relative afferent defect OD. Versions were smooth and full in all positions of gaze, with mild pain OD in superior gaze. Confrontation fields revealed a dense central scotoma with a superior and inferior nasal defect OD. Humphrey visual field testing confirmed an early superior arcuate defect with inferior depression OS and a dense superior and inferior arcuate defect with dense central scotoma OD. Color vision by Ishihara plates revealed 0/14 OD and 14/14 OS. Slit lamp examination was unremarkable OU. Intraocular pressures as measured by Goldmann tonometry were 15mmHg OU. Blood pressure was measured at 158/90 mmHg right arm sitting. Dilated fundus exam revealed pink, distinct, flat nerves with cup to disc ratios of .3/.3 OD and .45/.45 OS. No breaks or detachments were noted OU. Neurologic examination revealed cranial nerves V, VII - XII to be intact. Motor, sensory, and coordination testing were unremarkable. The patient was alert and oriented.

It was recommended that the patient undergo laboratory testing in the form of CBC with differential, ESR, c-reactive protein, platelet count, serum folate, vitamin B 12, methylmalonic acid, homocysteine, Lyme titer, ACE, ANA with reflex titer, RPR, FTA-ABS, SPEP, BUN and creatinine. Once kidney function was determined, she was sent for MRI of the brain and orbits.

Laboratory testing was remarkable for elevated homocysteine at 18.3, as well as possible monoclonal protein present on SPEP. MRI revealed enhancement of a long segment of the right optic nerve and questionable small white matter changes.

Upon examination five days later, the patient reported worsened vision OD, with continued pain upon eye movements. At this visit she reported possible bladder issues over the past year that had been attributed to diuretic side effects. Best-corrected visual acuity was hand motion OD and 20/20 OS. Pupils were equal and round, with a >1.8 log unit neutral density filter relative afferent papillary defect OD. Confrontation fields were consistent with hand motion OS. Humphrey visual field revealed stable defects OS and no remaining areas of preserved vision OD. Ocular motility testing demonstrated normal ductions, versions and saccades. Slit lamp examination was unremarkable OU. Intraocular pressures as measured by Goldmann tonometry were 16mmHg OD and 14mmHg OS. The right optic disc now exhibited early signs of papillitis. The left optic disc was stable without signs of edema.
3. Possible Causes of Unilateral Retrobulbar Optic Neuritis

Initial Considerations (before testing)
- Single and relapsing isolated optic neuritis
- Chronic relapsing inflammatory optic neuropathy
- Multiple Sclerosis
- Neuromyelitis Optica
- Acute disseminated encephalomyelitis
- Sarcoidosis
- Systemic Lupus Erythematosus
- Sjogren's syndrome
- Antiphospholipid antibody syndrome
- Behcet's Disease
- Wegener's granulomatosis
- Giant Cell Arteritis
- Lyme Disease
- Syphilis
- Primary Tumor
- Metastatic Tumor

Leading Considerations (after testing)
- Multiple Sclerosis
- Neuromyelitis Optica

4. Diagnosis

Clinical findings and MRI results obtained after initial examination confirmed retrobulbar optic neuritis as the cause of the patient's vision loss OD. Her year long history of finger paraesthesia and incontinence lead us to consider demyelinating disease as the cause of her condition.

Characteristics of NMO
- Characterized by severe attacks of optic neuritis
- Disproportionately strikes non-whites in developed nations
- Median age is 39 (decade older than median age of multiple sclerosis)
- MRI of the brain is usually normal
- Attacks are commonly unilateral
- Females are affected more often
- Visual prognosis is poor

- NMO diagnostic criteria (Wingerchuk et al)
  - Optic neuritis
  - Acute myelitis
  - At least two of the following
    - contiguous spinal cord MRI lesion which extends over at least 3 vertebral segments
    - brain MRI not supportive of MS
    - NMO IgG seropositive status (antibody to aquaporin 4)

5. Treatment/Management

The patient was admitted to the hospital for additional testing and appropriate treatment. MRI of the brain and cervical spine showed no definite abnormal signal to suggest demyelinating disease. The patient refused lumbar puncture, so further investigation into infectious and inflammatory processes within the cerebrospinal fluid was not possible. She was treated with
intravenous methylprednisolone, however her vision showed little improvement.

She returned for examination three days after she was discharged from the hospital. Her visual acuity was hand motion OD within her remaining temporal visual field and 20/20 OS. Color vision revealed 0/14 OD and 14/14 OS correct Ishihara plates. Pupils were isocoric with a >1.8 log unit neutral density filter relative afferent defect in the right eye. Humphrey visual field testing remained stable OS, with no remaining central field OD. Ocular motility demonstrated normal ductions, versions, and saccades, with mild pain on upgaze OD. Dilated fundus exam revealed an improved appearance of the disc OD, with no significant edema. This improved appearance of the nerve was best evaluated by comparing serial photographs taken at the two previous visits. We will continue to monitor her visual status and will consider NMO antibody (NMO-IgG) testing as well as MRI of the thoracic spine, if vision does not show significant improvement.

During acute episodes, both NMO and MS are treated with IV methylprednisolone. If NMO is confirmed, the diagnosis becomes important for relapse prevention. Prevention in NMO revolves around the use of immunosuppressive drugs whereas in MS, immunomodulatory drugs are used. Patients with NMO are typically treated with oral prednisolone with azathioprine (or mycophenolate mofetil), mitoxantrone, or monoclonal antibodies. This differs from the current treatment approach to MS which includes beta interferons, glatiramer, natalizumab as well as mitoxantrone.

6. Conclusion

It is important to consider neuromyelitis optica as a potential cause of optic neuritis. Although these patients receive the same initial treatment as multiple sclerosis related optic neuritis, the longer term treatment differs. In this case, neuromyelitis needs to be considered due to severe vision loss. If visual improvement does not occur as expected with typical optic neuritis secondary to MS, further investigation into possible NMO will need to be considered.

7. Bibliography


